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Left Recurrent Laryngeal Nerve Palsy Associated With Primary Pulmonary Hypertension and Patent Ductus Arteriosus

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Two patients with left recurrent laryngeal nerve paralysis in association with pulmonary artery hypertension are described. One had primary pulmonary hypertension and the other had patent ductus arteriosus. The greatly dilated pulmonary artery in these patients resulted in compression of the left recurrent laryngeal nerve and produced a cardiovocal (Ortner's) syndrome. The pathogenesis of the vocal cord palsy was documented by cross-sectional computed tomography.

In conclusion, computed tomography is of great help in differentiating this syndrome from other diseases such as mediastinal mass or lymphadenopathy whenever hoarseness is complicated by pulmonary hypertension.

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Paralysis of the left recurrent laryngeal nerve as a complication of various types of heart disease is a well known clinical entity. In 1897, Ortner (1) was the first to describe this syndrome in cases of mitral stenosis. Since then, many other investigators have reported Ortner's syndrome in patients with mitral stenosis, but the syndrome in association with primary pulmonary hypertension or patent ductus arteriosus is rare.

Case Reports

Case 1. A 34 year old woman was admitted to the hospital in August 1982 because of progressive exertional dyspnea and palpitation for the past 4 years. Her symptoms began at 30 years of age in the postpartum period of a second pregnancy and after the delivery of twins. Two years later, she developed chest pain at rest. Five months before admission, she developed dyspnea, even at rest, and intermittent hoarseness of her voice that soon became constant.

On examination, she was in sinus rhythm with a slight increase in jugular venous pressure. Blood pressure was 100/70 mm Hg. Her chest X-ray film showed enlargement of the heart and main pulmonary artery (Fig. 1A). The electrocardiogram showed a marked degree of right atrial overload and right ventricular hypertrophy (Fig. 2A). The pulse tracing in the second intercostal space and along the left sternal border showed pulmonary artery pulsation and a sustained right ventricular impulse, respectively. The pulmonary ejection sound and pulmonary second heart sound were increased in intensity in the second intercostal space (Fig. 3).

On echocardiography, the left ventricle was normal, but the right ventricle was dilated without paradoxical septal motion. The pulmonary valve showed a systolic notch, a flat EF slope and a decreased A wave (Fig. 4). Enhanced computed tomography of the chest showed enlarged pulmonary arteries without hilar lymph node swelling (Fig. 5A). Indirect laryngoscopy showed complete paralysis of the left vocal cord.

Cardiac catheterization data (Table 1) disclosed a pulmonary artery pressure of 86/36 mm Hg. Pulmonary angiography was not performed because of the severe pulmonary hypertension with dilated pulmonary arteries.

Case 2. A 57 year old woman was admitted in January 1983 for progressive dyspnea and palpitation on effort lasting for 10 years. At 47 years of age, she first experienced dyspnea and was hospitalized. The diagnosis of patent ductus arteriosus was made, and she had been taking diuretic drugs since then. In December 1982, she developed dyspnea at rest and hoarseness. A month later she was admitted to the hospital for further study.

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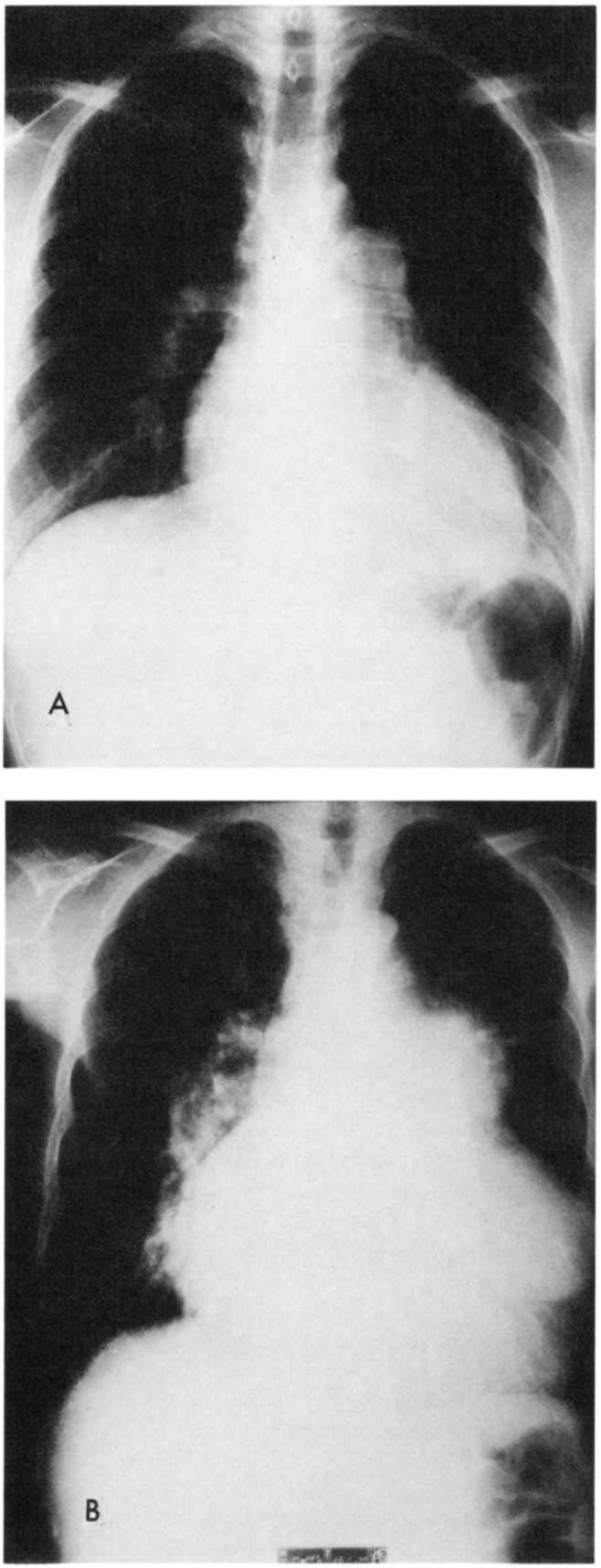

Figure 1. Chest X-ray films of Case 1 (**A**) and Case 2 (**B**). Both films show generalized cardiomegaly and enlargement of the main and left pulmonary arteries.

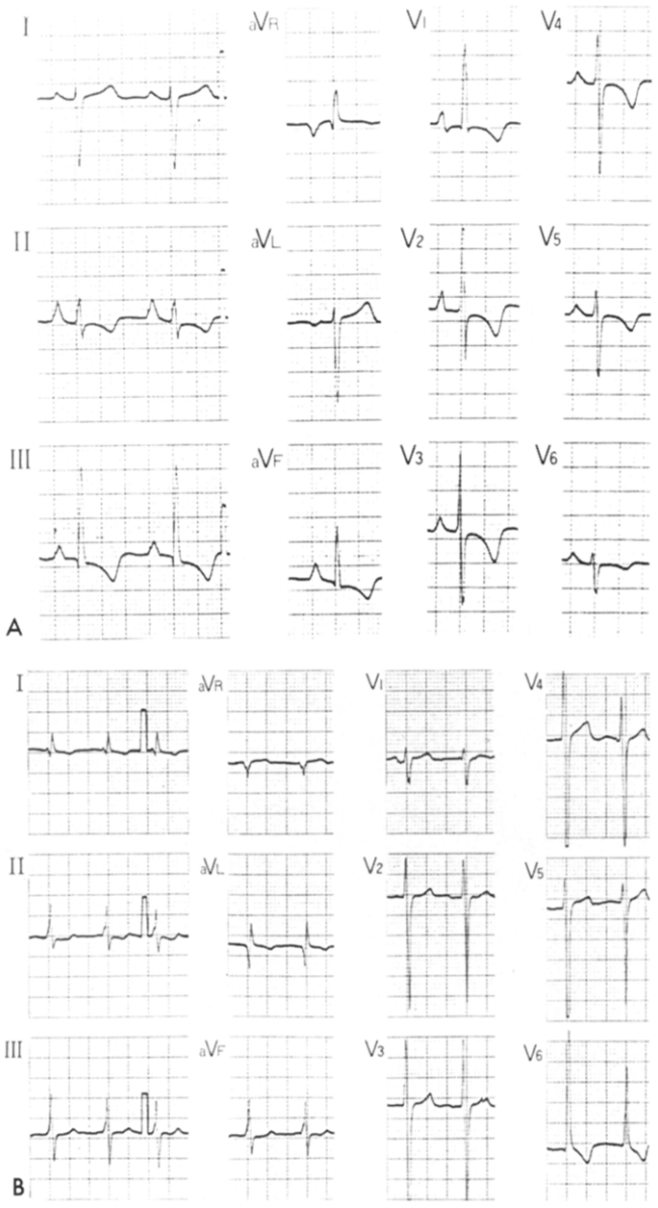

Figure 2. Electrocardiograms showing a marked degree of right atrial overload and right ventricular hypertrophy in Case 1 (**A**) and biventricular overload and atrial fibrillation in Case 2 (**B**).

On examination, she was in atrial fibrillation with a moderate increase in jugular venous pressure. Blood pressure was 120/60 mm Hg. She had a prominent right ventricular impulse and pedal edema. Auscultation over the apex showed a grade 3/6 holosystolic murmur and a prominent S_3 as well as a short mid-diastolic rumbling murmur. Over the second left intercostal space, the second heart sound was accentuated and closely split, and a grade 3/6 ejection systolic murmur and an early blowing diastolic murmur were heard. Over the third left intercostal space, a high-pitched diastolic murmur was audible. Over the left lower sternal border, a grade 2/6 pansystolic murmur that increased on inspiration

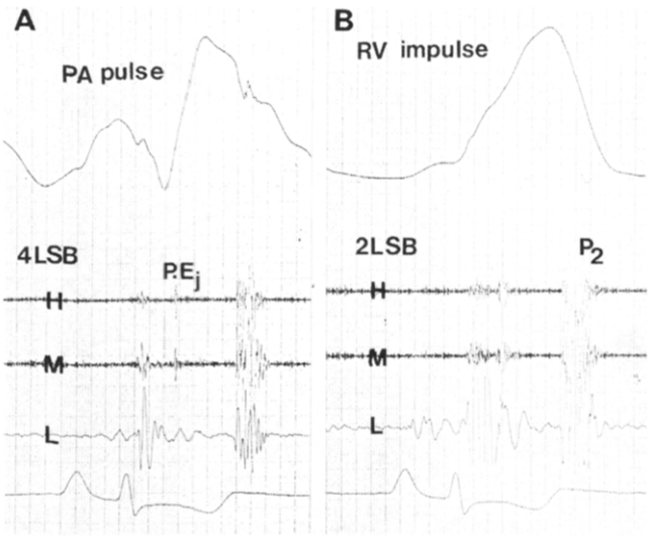

Figure 3. Pulse and phonocardiographic tracings. **A**, Case 1. The pulse tracing in the second intercostal space shows pulmonary artery (PA) pulsation. **B**, Case 2. The pulse tracing along the left sternal border shows a sustained right ventricular (RV) impulse with a large A wave. In both phonocardiograms, the pulmonary ejection sound (P.Ej) and pulmonary component of the second heart sound (P_2) are increased in intensity. H, M and L = high, medium and low frequency recordings, respectively; LSB = left sternal border.

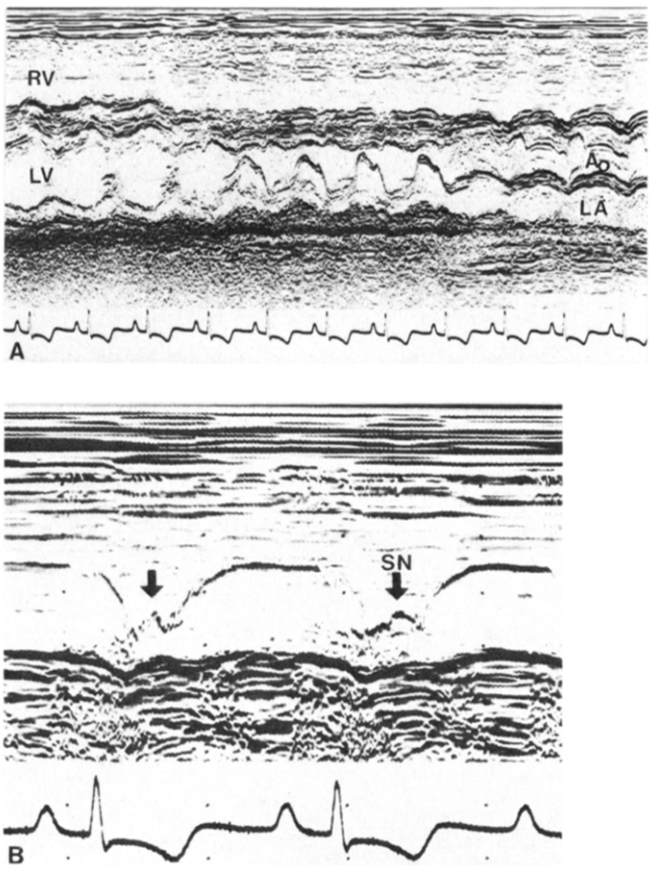

Figure 4. M-mode echocardiograms. **A**, Case 1. Recordings over the left sternal border. The sector scan from the left ventricle (LV) to the aortic (Ao) root shows dilation of the right ventricular (RV) cavity without paradoxical septal motion. **B**, Case 2. Pulmonary valve echocardiogram shows a systolic notch (SN) (**arrows**), flat EF slope and decreased A wave. LA = left atrium.

was also heard. There were a few rales over both lung fields posteriorly. The liver was moderately enlarged and pulsatile.

The chest X-ray film showed generalized cardiomegaly and enlarged main and left pulmonary arteries (Fig. 1B). The electrocardiogram showed biventricular overload and atrial fibrillation (Fig. 2B).

The echocardiogram showed large left and right ventricles without paradoxical septal motion. The pulmonary valve recording showed a typical pattern of pulmonary hypertension. Enhanced computed tomography of the chest demonstrated dilated pulmonary arteries with no enlarged hilar lymph nodes (Fig. 5B). Indirect laryngoscopy showed that the left vocal cord was in the midline position and was paralyzed.

After cardiac catheterization and angiography (Table 1), diagnosis was made of patent ductus arteriosus with pulmonary hypertension and no reversal of shunt, pulmonary regurgitation, tricuspid regurgitation and congestive heart failure.

Discussion

Ortner's syndrome (1), or the "cardiovocal syndrome," is the name originally given to paralysis of the left vocal cord in cases of mitral stenosis associated with a large left atrium and pulmonary artery dilation (2). However, this syndrome has been reported not only in cases of mitral valvular disease (1,3–7), but also in cases of left ventricular failure caused by hypertensive heart disease (8,9), atrial septal defect (10,11), Eisenmenger's syndrome (12), patent ductus arteriosus (10,13,14), primary pulmonary hypertension (14–19) and pulmonary embolism (19,20).

Causes of left laryngeal nerve palsy. The pathogenesis of the left recurrent laryngeal nerve paralysis in the cardiovocal syndrome is disputed. According to Ortner (1), an enlarged left atrium can directly compress the left recurrent laryngeal nerve against the aortic arch. Other investigations have attributed nerve injury to traction on the nerve by the aorta drawn caudally by a greatly hypertrophied right ventricle or compression of the nerve between the aorta and a dilated pulmonary artery (3,4) and to compression of the nerve between the aorta and greatly dilated ductus arteriosus (15).

The left recurrent laryngeal nerve originates on the anterior surface of the aortic arch and is immediately medial to the phrenic nerve, which is also crossing the aorta. As the nerve loops under the aorta, it is distal to the ligamentum arteriosum (or ductus arteriosus). A space between main pulmonary artery and aortic arch (aortic window) is present

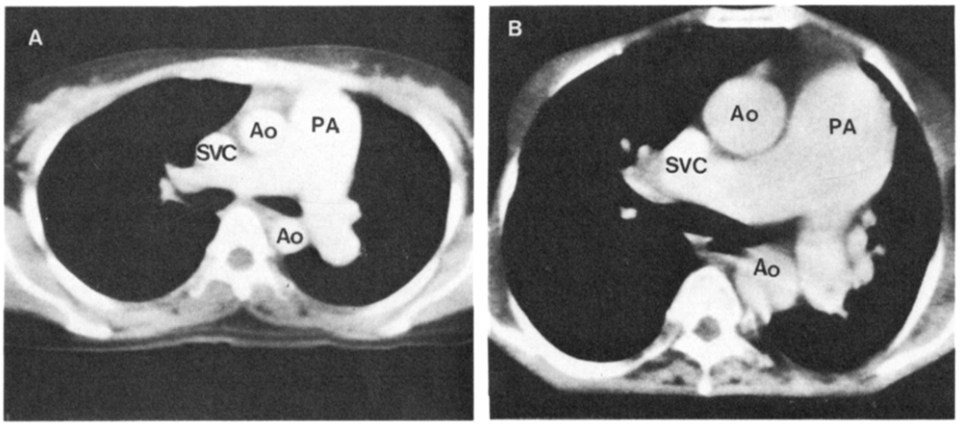

Figure 5. Enhanced computed tomographic scans of the chest. **A**, Case 1. **B**, Case 2. Both examinations show dilated pulmonary arteries without hilar lymph node swelling (see text). Ao = aorta; PA = pulmonary artery; SVC = superior vena cava.

in the anatomically normal individual (Fig. 6A). Therefore, the pulmonary truncus is not present in the tomographic cross-section through the lower border of the aortic arch, but is present at the carina level (21,22).

An enlarged pulmonary truncus in our two cases of Ortner's syndrome was adjacent to the lower border of the aortic arch because the pulmonary truncus protruded cephalad. Therefore, the pulmonary truncus and lower border of the aortic arch were present in the same tomographic section (Fig. 6B). It is suggested that compression of the nerve may be attributed to the abnormal anatomic relations between the enlarged pulmonary truncus and lower border of the aortic arch. This analysis of pathogenesis by cross-sectional computed tomographic anatomy has never been discussed. An enlarged pulmonary truncus resulting in compression of the left recurrent laryngeal nerve may be documented by computed tomography easily and noninvasively.

Our cases support the view that the nerve palsy in the cardiovocal syndrome can be caused by compression of the nerve by a dilated pulmonary artery and does not necessarily require an enlarged left atrium. It has been suggested that left vocal cord paralysis in heart disease may be taken as evidence of increased pulmonary artery pressure and an enlarged main pulmonary artery.

Implication. Visualization of the vocal cords by indirect laryngoscopy should be done routinely in the presence of heart disease with pulmonary hypertension. Computed tomography should be of great help in differentiating this

Table 1. Catheterization Data

	Case 1		Case 2	
Position	Pressure (mm Hg)	O_2 Saturation (%)	Pressure (mm Hg)	O_2 Saturation (%)
PA				
Wedge	($\bar{5}$)	74	($\bar{25}$)	88
Main	86/36 ($\bar{50}$)	60	72/42 ($\bar{52}$)	77
RV				
Outflow		60.5		74
Apex	86/00 (16,edp)	61.5	72/8 (20,edp)	56
Inflow		61		61
RA				
High		61.5		54
Middle	($\bar{6}$)	61.5	($\bar{19}$)	54
Low		64		57
SVC	($\bar{7}$)	63.5	($\bar{20}$)	51
IVC	($\bar{7}$)	72	($\bar{20}$)	53
Ao				
Ascending			108/64 ($\bar{80}$)	87
Descending		75	112/52 ($\bar{72}$)	86
			L→R shunt	62

Ao = aorta; edp = end-diastolic pressure; IVC = inferior vena cava; L→R = left to right; O_2 = oxygen; PA = pulmonary artery; RA = right atrium; RV = right ventricle; SVC = superior vena cava. Numbers in parentheses with overbar indicate mean pressures.

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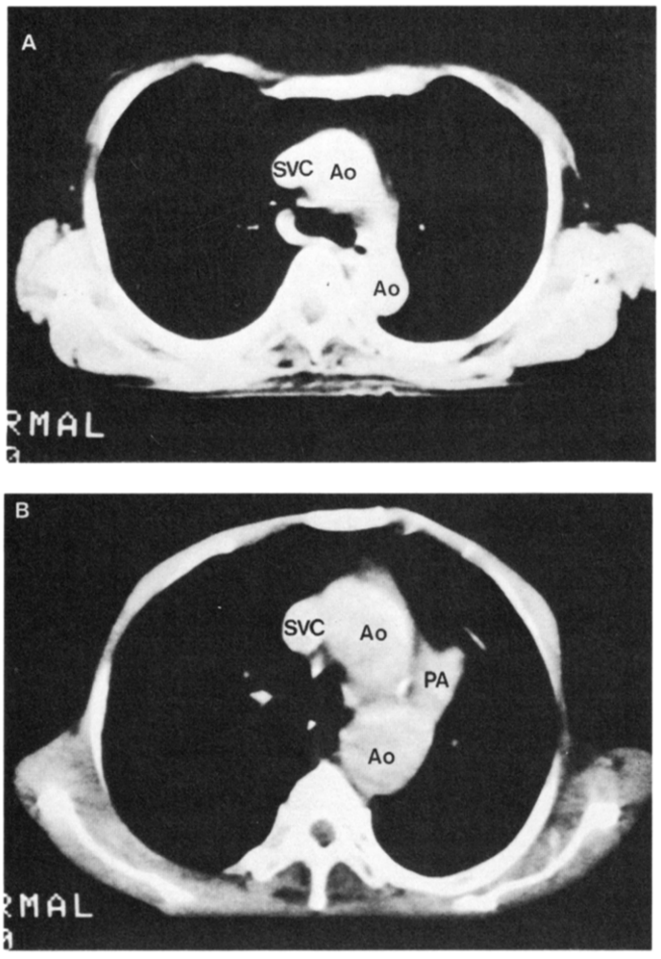

Figure 6. Enhanced computed tomographic cross-sectional scans through the lower border of the aortic arch in a normal anatomic case (**A**) and in Case 2 (**B**). The main and left pulmonary arteries are not present at this level in the normal case, but are present in the same section in Case 2. Ao = aorta; PA = pulmonary artery; SVC = superior vena cava.

syndrome from other disorders such as mediastinal mass or lymphadenopathy whenever hoarseness is complicated by pulmonary hypertension.

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